International Journal of Surgery Case Reports 4 (2013) 822–824

International Journal of Surgery Case Reports

journal homepage: www.casereports.com

Primary paraganglioma of seminal vesicle

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ARTICLE INFO

Article history:
Received 2 July 2013
Received in revised form 16 July 2013
Accepted 19 July 2013
Available online 26 July 2013

Keywords:
Seminal vesicle
Seminal vesicle tumor
Paraganglioma

ABSTRACT

INTRODUCTION: Paragangliomas are rare tumors arising from neural crest tissue located outside the adrenal gland. Primary seminal vesicle paraganglioma is extremely rare entity.
PRESENTATION OF CASE: A 26-year-old male patient presented with symptoms and signs of acute appendicitis where a CT of abdomen and pelvis showed an inflamed appendix and incidental finding of left seminal vesicle mass. The patient underwent uneventful laparoscopic appendectomy followed by transrectal ultrasound (TRUS) guided seminal vesicle biopsies. Histopathology revealed a neuroendocrine neoplasm consistent with paraganglioma. Surgical excision of the left seminal vesicle was carried out.
DISCUSSION: Paraganglioma of genitourinary tract is rare. The urinary bladder is the most common site, followed by the urethra, pelvis and ureter. Seminal vesicle paragangliomas were reported in association with other genitourinary organ involvement such as bladder and prostate. Isolated seminal vesicle paraganglioma is extremely rare and surgical excision remains the standard treatment for localized paraganglioma.
CONCLUSION: Primary tumors of seminal vesicle are rare and represent a diagnostic challenge. Differential diagnosis includes a list of benign and malignant tumors. Primary seminal vesicle paraganglioma is a rare but important diagnosis to be included in the differential diagnosis.

1. Introduction

Extra-adrenal paragangliomas are tumors that arise from neural crest cells or organs. They are most frequently found in the organ of Zuckerkandl, but can be found anywhere along the sympathetic chain. Generally paraganglioma are slow growing benign tumors, although malignant degeneration and distant metastases has been described. Seminal vesicle paragangliomas have been reported in association with bladder and prostate involvement, but as primary seminal vesicle paraganglioma there was only one case reported in medical literature to the best of our knowledge.

2. Case report

A 26-year-old male patient with unremarkable past medical and surgical history, presented to accident and emergency department with symptoms and signs of acute appendicitis where a CT with contrast of abdomen and pelvis showed an inflamed appendix and incidental finding of left enhancing seminal vesicle mass (Fig. 1). The patient underwent uneventful laparoscopic appendectomy then referred to our care. A transrectal ultrasound (TRUS) guided seminal vesicle biopsies was done and the patient did not experience any symptoms or signs of catecholamine overproduction. Histopathology and immunohistochemistry revealed a neuroendocrine neoplasm most consistent with paraganglioma. The tumor cells stain strongly positive with chromogranin and synaptophysin. They show strong cytoplasmic staining for Myo-D1 (non-specific) and weak cytoplasmic staining for CD117. S100 highlights the sustentacular cells around the cellular nests. Inhibin, HMB-45, CD31, desmin, SMA, CKAE1/AE3, CD34 and calretinin were negative.

Work up for functional paraganglioma was carried out; the 24-h urinary catacholamine metabolites were within the normal range. The patient then underwent surgical exploration and excision of the left seminal vesicle. Macroscopically, the seminal vesicle was brownish in color with soft to firm in consistency, measuring $2 \times 2 \times 1.3$ cm, and weighing 8.80 g. Microscopically, nests of cuboidal cells separated by vascular fibrous septa without evidence of vascular invasion, mitotic figures or necrosis. The surgical margins were free from tumor. A panel of immunohistochemistry was done. Chromogranin, synaptophysin and CD56 are positive in tumor cells and S100 was positive in sustentacular cells. These findings confirm the diagnosis of paraganglioma. Follow-up imaging with CT scan done one year after surgery showed no evidence of local recurrence or distant metastasis (Fig. 2).

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http://dx.doi.org/10.1016/j.ijscr.2013.07.009

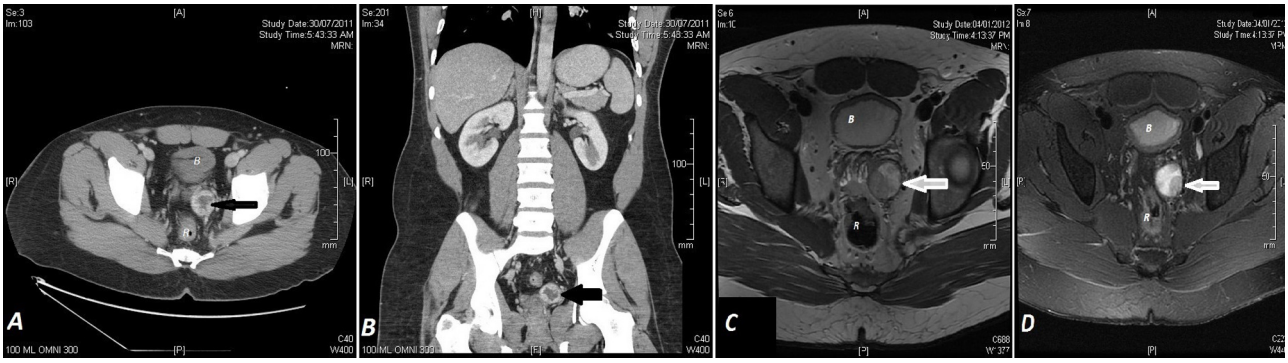

Fig. 1. CT (A and B) and MRI (C and D) of abdomen and pelvis: showing a well-defined rounded left SV (arrow) enhancing necrotic mass measuring 2.7 × 2.7 × 2.9 cm. (D) The lesion appears hyperintense on T2 and shows significant enhancement on the post contrast images mainly at its periphery. No regional lymphadenopathy or invasion of the adjacent structures. Bladder (B) and rectum (R).

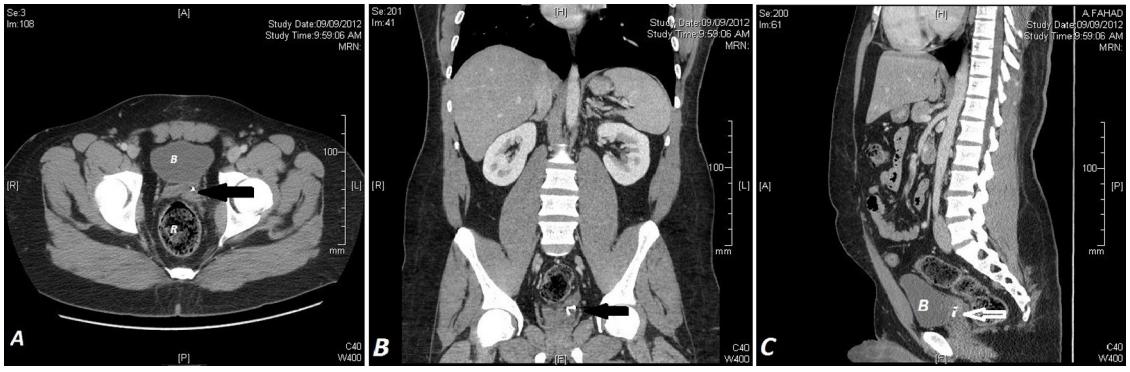

Fig. 2. Follow-up CT scan of abdomen and pelvis one year post-operatively showing surgical clips (arrow) at the site of left SV with no evidence of local recurrence or distant metastasis. Bladder (B) and rectum (R).

3. Discussion

Extra-adrenal paragangliomas are rare neuroendocrine tumors that arise from neural crest-derived endocrine cells anywhere along the sympathetic chain. Nearly 85% of paragangliomas are intra-abdominal, 12% are intrathoracic, and 3% are cervical. Paragangliomas may occur at unusual locations, including the kidneys, urethra, urinary bladder, prostate, spermatic cord, gallbladder, uterus and vagina.[3] In the genitourinary tract, the urinary bladder is the most common site for paraganglioma (79.2%), followed by the urethra (12.7%), pelvis (4.9%), and ureter (3.2%).[10,11] Furthermore, approximately 10% of all extra adrenal paraganglioma are malignant.[11] The histogenesis of paragangliomas of the spermatic cord is unknown, although it has been speculated that paraganglion nests in the spermatic cord may be secondary to dysgenesis during embryogenesis.[3] The origin of seminal vesicle paraganglioma can be explained in the same way as for the spermatic cord, because the cells that give rise to the seminal vesicle originate in the caudal Wolffian duct and urogenital sinus.[1,3] Histologically, paragangliomas show similar morphologic characteristics. They exhibit a pattern of cell clusters (Zellballen) surrounded by fine fibrovascular septa. Immunohistochemistry for neuroendocrine markers are positive for synaptophysin, chromogranin A, and CD 56. Protein S100 highlights the sustentacular and tumor cells.[13] The distinction between benign and malignant paraganglioma represent a diagnostic challenge. Clinical parameters proposed to predict malignancy include higher urinary catecholamine metabolites such as VMA and dopamine, an extra-adrenal tumor location, large tumor size, and persistent postoperative hypertension.[2] Histopathological findings of tumor necrosis, vascular invasion, capsular invasion and high mitotic figures indicate malignancy. The only currently accepted mean to define malignant paraganglioma is the presence of metastatic deposits at non-chromaffin sites.[5] Currently, there is no standard staging system for paraganglioma. The clinical presentation of paragangliomas is highly variable, largely depending on the size and location of the lesion, as well as the functional status of the tumor. If the tumor is functional, the patient may experience symptoms and signs of catecholamine overproduction, such as hypertension, headaches, palpitations, sweating, tachycardia and anxiety, and may be precipitated by an inciting event depending on its location such as micturition in case of bladder paraganglioma. The laboratory diagnosis of functional paragangliomas is made by detecting an elevation of the 24-h urinary of catecholamine metabolites and vanillylmandelic acid levels.[4,6–9] Whereas in non-functional tumors, the diagnosis may not be made until an advanced stage due to asymptomatic presentation or nonspecific symptoms or as an incidentloma as in our case. Treatment options of paraganglioma include catecholamine blockade, surgery, chemotherapy, radiofrequency ablation, cryoablation and radiation therapy. The standard treatment for localized or locally advanced paraganglioma is surgery while unresectable or metastatic tumors are treated with palliative therapy.[12]

4. Conclusions

Extra-adrenal paragangliomas are rare neuroendocrine tumors. In the genitourinary tract, the urinary bladder is the most common site for paraganglioma. Primary seminal vesicle paraganglioma

is extremely rare with only one reported case in literature. A second case of primary seminal vesicle paraganglioma presented.

Conflict of interest

No conflicting interests.

Funding

None.

Ethical approval

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in Chief of this journal on request.

Author contributions

Study design, Data collections, Data analysis, Writing done by Dr. Badr Alharbi.

Review was done by Dr. Abdullah Alghamdi.

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